

CONCLUSIONS: Screening and follow-up of newborns for SCD is feasible in a developing country in Africa. Extra effort in tracking is necessary to ensure that infants with disease are found early and referred for medical management.

PROSPECTIVE FOLLOW-UP OF PATIENTS WITH EWING SARCOMA WITHIN THE LATE EFFECTS SURVEILLANCE SYSTEM

Submitted by Marios Paulides

Marios Paulides^a, Uta Dirksen^b, Wolfgang Stohr^a, Heribert Jürgens^b, Helmuth-Gunther Dorr^a, Tobias Bolling^c, Normann Willich^c, Jorn-Dirk Beck^a, Thorsten Langer^a

^aLESS Study Center, University Hospital for Children and Adolescents, Erlangen, Germany; ^bDepartment of Pediatric Hematology and Oncology, Ewing's Sarcoma Trial Center, and ^cDepartment of Radiotherapy, RISK Study Center, University Hospital Muenster, Muenster, Germany

INTRODUCTION: It is known that antineoplastic treatment may induce early and late organ toxicities depending on treatment modalities and intensity.

OBJECTIVE: The aim of this study was to determine the cumulative incidence of sequelae within our cohort of patients treated within the EICESS-92 (European Intergroup Cooperative Ewing's Sarcoma Study, 1992) treatment trial.

METHODS: Since 1998, the Late Effects Surveillance System (LESS) of the German Society for Pediatric Oncology and Hematology has prospectively registered late effects in patients of all ages with relapse-free bone and soft tissue sarcoma in Austria, Germany, and Switzerland. The follow-up is conducted locally in accordance with LESS guidelines. Data are reported to the LESS center for collation and analysis.

RESULTS: There were 67 patients available for analysis (42 male, 25 female) with a median age at diagnosis of 13 years and a median follow-up of 3.5 years. Registration had to be terminated for 17 patients as a result of relapse. In total, 43.3% (29 of 67) of the patients were reported to have at least 1 sequelae of treatment. Sixteen patients suffered toxicity in 1 organ system, 9 patients developed toxicity in 2 organ systems, and there were 3 organ systems affected in 4 patients. Nephrotoxicity was reported in 10.4% (7 of 67), cardiotoxicity in 8.9% (6 of 67), peripheral polyneuropathy in 5.9% (4 of 67), and other toxicities in 34.3% (23 of 67) of the patients.

CONCLUSIONS: Sequelae of treatment for Ewing sarcoma within this cohort of the EICESS-92 study were not more frequent than reported previously. Patients are at risk for the development of several toxicities after treatment for Ewing sarcoma, and they should receive adequate medical follow-up.

HEMATOPOIETIC STEM CELL TRANSPLANTATION FOR THE TREATMENT OF THALASSEMIA: THE GREEK EXPERIENCE

Submitted by Ioulia Peristeri

Ioulia Peristeri, Vassiliki Kitra, Evgenios Goussetis, Georgios Vessalas, Maria Theodosaki, Anna Paisiou, Eftychia Petrakou, Stelios Graphakos
Bone Marrow Transplantation Unit, Agia Sophia Children's Hospital, Athens, Greece

INTRODUCTION: Although prevention remains the cornerstone for the management of thalassemia, hematopoietic stem cell transplantation (HSCT) is the only curative approach.

OBJECTIVE: Our goal was to assess our experience with HSCTs for the treatment of patients with thalassemia.

METHODS: From 1994 to 2006, 96 HSCTs have been performed in 84 thalassemic children from Greece, 3 with sickle cell/thalassemia and 1 with sickle cell disease. According to Pesaro classification, of these 84 children, 20 were in class I, 35 were in class II, and 29 were in class III. Donors were 84 histocompatible siblings and 4 unrelated volunteers. The graft was of bone marrow in 85, cord blood in 3, bone marrow and cord blood in 4, and peripheral blood stem cells in 4. The conditioning regimen consisted of busulfan, cyclophosphamide, and antilymphocyte globulin.

RESULTS: All except 1 patient received engraftments. Ten patients rejected the graft. Eight received another transplant from the same donor, 7 of which were successful. Four patients died; causes of death were graft-versus-host disease (GVHD) (2), disseminated toxoplasmosis (1), and brain hemorrhage (1). At a median follow-up time of 6.5 years, 84 of 88 children survived, 81 were cured and free from transfusions, and 3 remained transfusion-dependent. Severe acute GVHD developed in 18 children, and chronic GVHD developed in 8 patients. The overall survival rate, event-free survival rate, rejection rate, and transplant-related mortality rate were 95%, 94%, 11%, and 5%, respectively. Event-free survival was 100% for class I, 95% for class II, and 87% for class III. Eleven children had mixed chimera (residual recipient hematopoiesis) with normal levels of hemoglobin.

CONCLUSIONS: HSCT is a highly effective treatment for thalassemic patients who have a fully matched donor (related or unrelated). Younger age at transplant secures excellent results with reduced morbidity and mortality rates.

TWO-DECADE EXPERIENCE AND LONG-TERM SURVIVAL IN PEDIATRIC NON-HODGKIN'S LYMPHOMA

Submitted by Sophia Polychronopoulou

Vassilios Papadakis^a, Agapi Parcharidou^a, Anna Paisiou^a, Natalia Tourkantoni^a, Sofia Papargyri^a, Kalliopi Stefanaki^b, Sophia Polychronopoulou^a
Departments of ^aPediatric Hematology-Oncology and ^bDepartment of Pathology, Agia Sophia Children's Hospital, Athens, Greece

INTRODUCTION: Treatment results for pediatric non-Hodgkin's lymphoma (NHL) continue to improve internationally.

OBJECTIVE: Our goal was to evaluate patient characteristics in our series of patients with NHL and outcomes for the last 16 years (1990–2006).

METHODS: Our patients included 52 newly diagnosed children (11 girls) with a median age of 8.40 years (range: 0.33–14.5 years). Histology results included B-lymphocyte NHL, T-lymphocyte NHL, and Ki-1 in 35, 12, and 5 patients, respectively. In each 5-year period, 14 (3), 17 (3), and 21 (5) patients (girls) were diagnosed, respectively. Common presenting sites were the mediastinum (16), neck area (14), and abdomen (10). Disease was at stage I, II, III, and IV in 3, 14, 23, and 7 patients, respectively. Treatment varied over time. Berlin-Frankfurt-Munich (BFM) protocols had been applied since 1995 (BFM-NHL-90), and since 1997 the BFM-NHL-95 protocol had been applied. Irradiation was given to 5 patients (2 with B-NHL, 3 with T-NHL), and autologous stem cell transplantation was performed on 4 patients, all with B-NHL (1 with central nervous system disease, 1 with residual disease at the end of treatment, and 2 at relapse).

RESULTS: At this writing, 41 patients are alive; 39, 2, and 1 are in first, second, and third remission, respectively. In total, 9 have succumbed (2 died soon after admission in other hospitals as a result of acute-phase complications), and 5 patients died during the first decade of our retrospective study (with T-histology and extensive disease). The event-free survival rate is 74.4% (39 of 52 patients), and the overall survival rate is 80.9% (41 of 52 patients), for a median follow-up time of 6.1 years (range: 0.01–14.7 years) for all patients. For the 39 patients treated with the BFM-95 protocol since 1997, event-free survival and overall survival rates are 79.4% and 88.2%, respectively, for a median follow-up time of 4.8 years.

CONCLUSIONS: Overall and event-free survival rates and outcome of our patients with NHL treated during the last 16 years are standing high. There has been limited use of irradiation and stem cell transplantation.

SERUM LEVELS OF MYCN AMPLIFICATION IN NEUROBLASTOMA WITH TUMOR-RELEASED DNA BY REAL-TIME QUANTITATIVE POLYMERASE CHAIN REACTION

Submitted by Tohru Sugimoto

Tohru Sugimoto^a, Takahiro Gotoh^a, Tomoko Iehara^a, Kunihiko Tsuchiya^a, Miki Ohira^b, Akira Nakagawara^b, Hajime Hosoi^a
^aKyoto Prefectural University of Medicine, Kyoto, Japan; ^bChiba Cancer Center Research Institute, Chiba, Japan

INTRODUCTION: *MYCN* amplification (MNA) indicates a poor prognosis in neuroblastoma and is routinely assayed for therapy stratification.

OBJECTIVE: We aimed to develop a diagnostic tool to predict *MYCN* status by using serum DNA, which, in patients with cancer, predominantly originates from tumor-released DNA.

METHODS: Using DNA-based real-time quantitative polymerase chain reaction, we simultaneously quantified *MYCN* (2p24) and a reference gene, *NAGK* (2p12), and evaluated *MYCN* copy number as a *MYCN/NAGK* ratio in 87 patients with neuroblastoma whose *MYCN* status had been determined by Southern blotting. Of these patients, 17 had *MYCN*-amplified neuroblastoma, and 70 had nonamplified neuroblastoma.

RESULTS: The serum *MYCN/NAGK* ratio in the MNA group (median: 199.32; range: 17.1–901.6 [99% confidence interval: 107.0–528.7]) was significantly ($P < .001$) higher than that in the non-MNA group (median: 0.87; range: 0.25–4.6 [99% confidence interval: 0.82–1.26], Mann-Whitney U test). The sensitivity and specificity of the serum *MYCN/NAGK* ratio as a diagnostic test were both 100% when the serum *MYCN/NAGK* ratio cutoff was set at 10.0. Among 6 patients in the MNA group whose clinical courses were followed, the serum ratios decreased to within the normal range in the patients in remission ($n = 3$), but they rose to high levels in the patients who had a relapse ($n = 2$) or failed to achieve remission ($n = 1$). The serum *MYCN/NAGK* ratio in the MNA group is likely to be the more sensitive tumor marker than conventional urinary vanillylmandelic and homovanillic acid markers and neuron-specific enolase markers to predict patients' clinical course.

CONCLUSIONS: Measurement of the serum *MYCN/NAGK* ratio seems to be a promising method for accurately assessing *MYCN* status in neuroblastoma.

RECURRENT IDIOPATHIC THROMBOCYTOPENIC PURPURA IN CHILDHOOD

Submitted by Maria Vranou

Maria Vranou, Helen Pergantou, Helen Platokouki, Dimitris Kousiades, Maria Avgeri, Sophia Aronis
Hemostasis Unit, Agia Sophia Children's Hospital, Athens, Greece

INTRODUCTION: Idiopathic thrombocytopenic purpura (ITP) is usually a benign disease that remits within

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